Functional and esthetic rehabilitation in amelogenesis imperfecta with all-ceramic restorations: A case report

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Amelogenesis imperfecta is a hereditary condition resulting in poor tooth development, severe anomalies, or complete absence of enamel. Enamel lesions may be the only characteristic of this condition or may be part of a generalized syndrome. Amelogenesis imperfecta can be characterized by enamel hypoplasia and/or hypomaturation or hypocalcification of the existing teeth. Restoration for patients with this condition should be oriented toward the functional and esthetic rehabilitation and the protection of the existing teeth. This paper presents a description of a patient's oral rehabilitation with all-ceramic veneers and crowns after extensive crown lengthening. The diagnostic procedure is also reported in detail. A microscopic examination was also performed on an extracted third molar under polarized light. *(Quintessence Int 2005;36:329–338)*

Key words: all-ceramic restorations, amelogenesis imperfecta

Amelogenesis imperfecta is a hereditary condition resulting in poor tooth development, severe enamel anomalies, or complete absence of tooth enamel. This condition is caused by improper differentiation of the ameloblast cells.[1] The genetic defects of the enamel are the most frequent congenital anomalies of the hard dental tissues. The enamel lesions may be the only characteristic of the whole condition, or they can accompany other lesions located in the dentin, or be part of generalized genetic disorders. The later group is termed as hypoplasia, hypomineralization and hypomaturation of the enamel, according to the clinical findings. Enamel anomalies are present in more than 70 syndromes.[2]

The hereditary disorders of enamel formation effect all the teeth in both primary and secondary dentitions. The most common form of amelogenesis imperfecta is the autosomal dominant hypocalcified type, followed by the hypomaturation and the hypoplastic type.[3–5]

Hereditary anomalies of enamel formation result in various forms of amelogenesis imperfecta that can be familial and inherited as autosomal dominant, autosomal recessive, or x-linked dominant.[6,7]

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Frequency

The frequency of amelogenesis imperfecta may vary among different populations or countries. In the United States, the frequency of all types was 1:14,000 and the most common type was autosomal dominant hypocalcified.[6] In Israel, the frequency was 1:8,000 and hypoplastic[8] was the most frequent type. In Sweden, the frequency was 1:4,000 and the hypoplastic type was the most common one. In an isolated region of this country the frequency was found to be 1:789.[9,10]

Classification

Amelogenesis imperfecta is subdivided into 14 different types, according to clinical appearance and inheritance patterns.[11] Clinical findings in this disorder, as well as in the enamel defects accompanying generalized genetic diseases, may vary according to the period which amelogenesis was effected by the genetic defect.[11–13] The four main types of amelogenesis imperfecta are: type I hypoplastic enamel, type II hypomaturated enamel, type III hypocalcified enamel, and type IV hypomaturated-hypoplastic enamel with taurodontism.[11]

Clinical findings

The clinical findings in amelogenesis imperfecta vary considerably among the different types.[11,12,14]

In type I, the lesions may appear as pits on the enamel surface of the tooth, or the teeth can be severely worn, without mesial or distal contact points. In these cases the enamel has a "snow-like" appearance. The enamel layer is usually thin and has a yellow-brown color. There are difficulties in tooth eruption and some teeth may erupt after root absorption has occurred. Cases of multiple missing permanent teeth have been reported.[4,15]

In type II, the enamel is hypomaturated and clinically has an opaque appearance. The enamel layer is normal in thickness, but softer than normal, and can be easily detached from the underlying dentin. Type III (hypocalcified enamel) is distinguished by severely worn teeth, as the enamel is detached from dentin within a short time after eruption. Teeth are very sensitive to thermal changes and are dark brown in color. Massive deposits and calculus are normally found around the teeth due to extreme sensitivity patients experience when they perform oral hygiene procedures.

Type IV is a combination of hypomaturated and hypoplastic enamel accompanied by taurodontism.[16–18] Many patients with amelogenesis imperfecta also exhibit open bite.[19]

Radiological findings

The radiological findings in patients with amelogenesis imperfecta include congenitally missing teeth, delayed dental eruption, crown resorption, root resorption, and pulp calcification on both erupted and unerupted teeth.

The density of the enamel layer in amelogenesis imperfecta seems lower than in normal teeth, and this finding is more pronounced in the hypocalcified type. Hypoplastic enamel shows great variation in density, and it may be difficult to distinguish from underlying dentin.[20]

The aim of this article is to describe the diagnostic procedure and the clinical stages in the rehabilitation, using all-ceramic veneers and crowns, of a patient with amelogenesis imperfecta.

CASE REPORT

Medical record

A 24-year-old patient was referred to the Postgraduate Clinic of the Department of Prosthodontics at the University of Athens for treatment. The patient reported he was suffering from Usher syndrome, which is characterized by reduced hearing ability (partial deafness) and problems in the eyes (pigmented retinopathy, retinitis pigmentosa). Usher syndrome is inherited in the autosomal recessive manner and is not related to amelogenesis imperfecta.[21]

Examination

The extraoral examination did not reveal anything significant. The intraoral examination revealed generalized enamel hypoplasia, which was more intense on the cusps of the posterior teeth and on the labial surfaces of

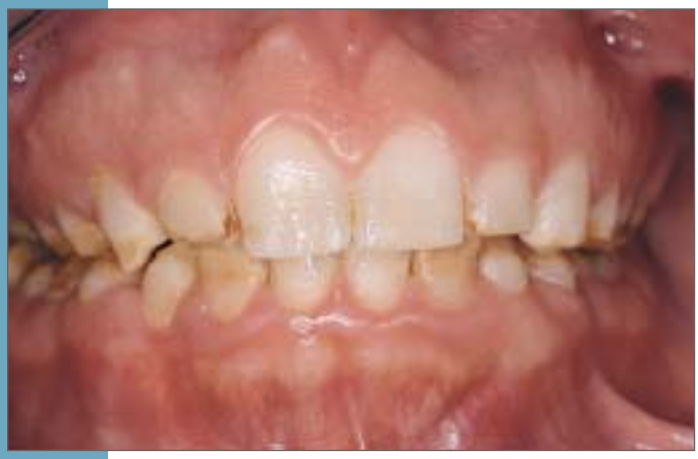


Fig 1 Initial clinical situation of the patient with amelogenesis imperfecta. The hypoplastic regions are mainly situated on the cervical third and on the occlusal surfaces.

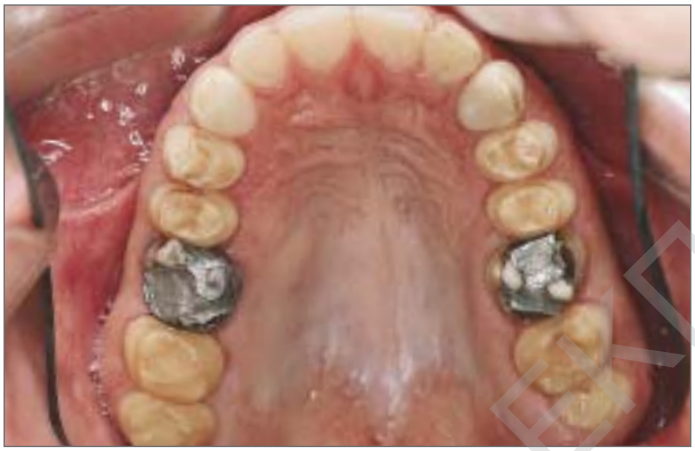


Fig 2 Occlusal view of the maxillary teeth.

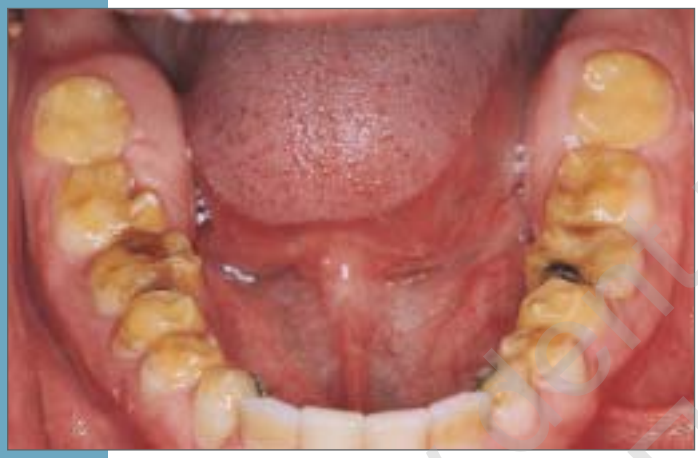


Fig 3 Occlusal view of the mandibular teeth.

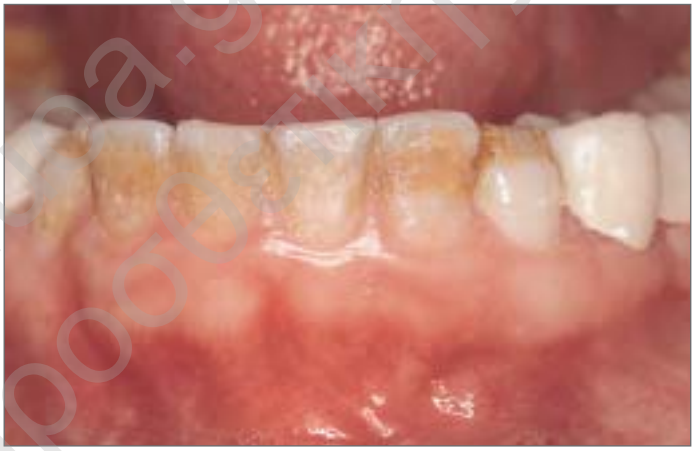


Fig 4 The mandibular anterior teeth with hypoplastic regions on the labial surfaces.

the mandibular anterior teeth. The enamel lesions on the maxillary incisors were limited to the incisal third (Figs 1 to 4).

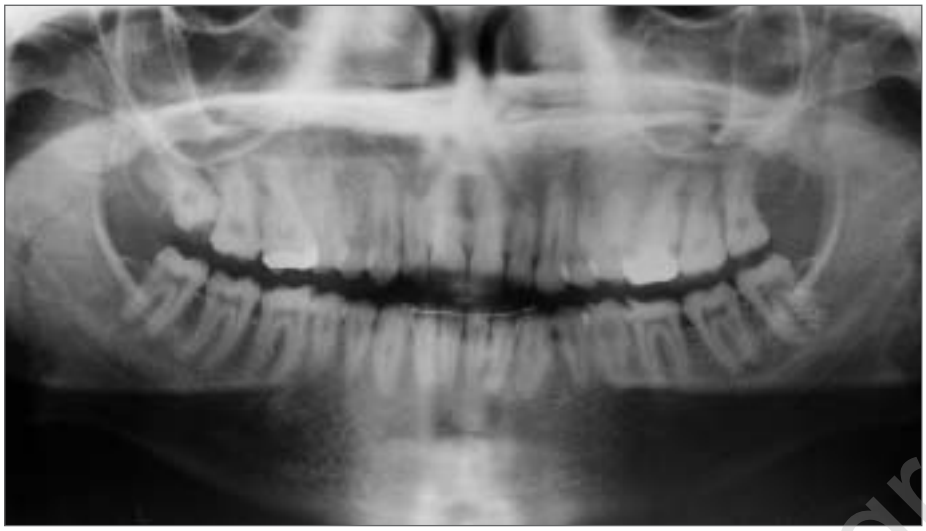
There were amalgam fillings on teeth 3 (16) and 14 (26), which showed increased marginal space around the amalgam margins. Tooth 3 (16) was endodontically treated. There were fractures on all occlusal cusps of the posterior teeth.

The irregular enamel surfaces were hard to examine with the dental probe and did not show any sign of detachment from the underlying dentin. The patient showed Class I occlusion with no discrepancy of the midline. The crown height was reduced (mainly on the posterior teeth), and the vertical dimension was slightly reduced (approximately 2 mm) due to tooth wear. The posterior teeth showed occlusal surface contacts instead of point contacts because of fractures. Cross-bite occlusion was noted on the right side.

The patient reported reduced chewing ability, and slight sensitivity to hot and cold food, and was not satisfied with the esthetic appearance of his teeth. The patient was also worried about the long-term prognosis of his teeth. There was slight inflammation on the gingival margins without any periodontal pockets. The gingival index was 40% and the oral hygiene index was 50%.

Radiological examination

The radiological examination with panoramic (Fig 5) and periapical x-rays did not reveal any missing teeth or periapical lesions. There



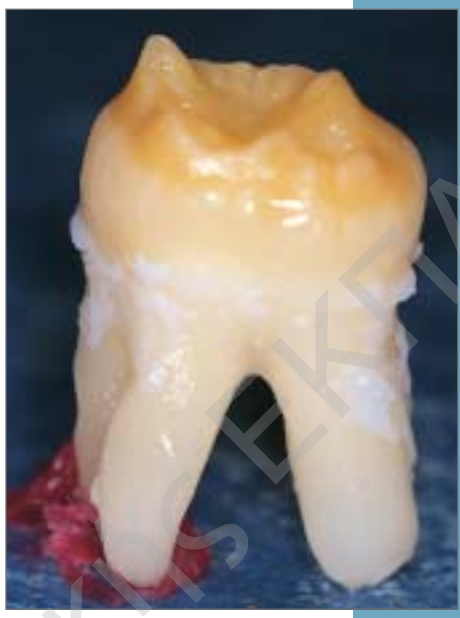


Fig 5 *(above)* Panoramic radiograph of the patient.
Fig 6 *(right)* The extracted tooth 1 (18). The enamel is irregular in shape, colored dark yellow-brown and hypoplastic.

was no bone loss. Teeth 1 (18), 16 (28), 17 (38), and 32 (48) had erupted. The pulp chambers of the teeth were regular in size. The enamel layer could be distinguished from the underlying dentin, but did not show the regular thickness and density.

Diagnosis

The clinical characteristics indicated amelogenesis imperfecta type I (hypoplasia) with "snow-like" enamel which was colored and appeared dark yellow or brown.

Microscopic examination

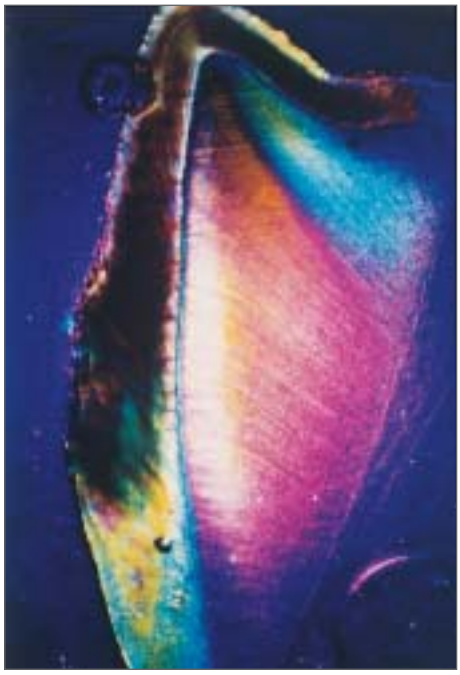
The maxillary third molar, tooth 1 (18), was extracted and kept in a 0.9% NaCl solution (Fig 6). The tooth was sectioned and examined with an optical microscope under polarized light (Figs 7 and 8) to verify the clinical diagnosis. The examination showed a thin hypoplastic enamel layer, and in some areas the dentin was exposed. The irregular surface was attributed to enamel hypoplasia, as this tooth was in the mouth for a shorter period than the other molars and was not fully functional. The enamel-dentin junction was physiological in the microscopic examination. The cementoenamel junction showed irregularities compared to a physiological tooth (Figs 8 and 9). According to microscopic examination, the amelogenesis in this patient could be classified as hypoplastic type I according to Darling's classification.[22]

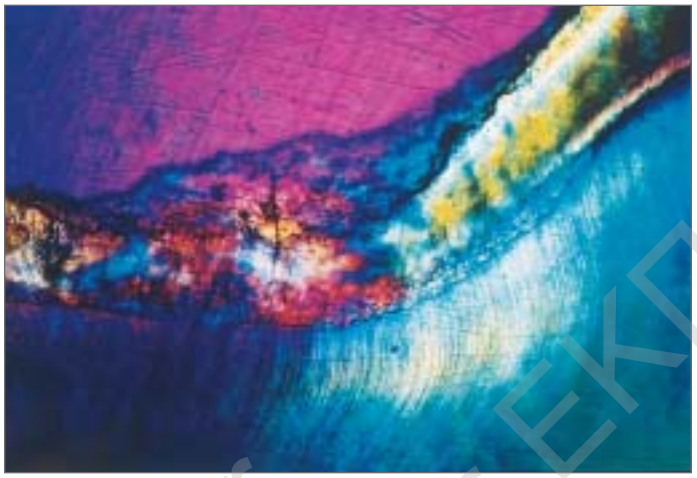
Treatment planning

Initial impressions were obtained from the patient, and study casts were constructed with hard stone. The study casts were mounted on a semi-adjustable articulator in centric relation, using a central relation intraoral registration. The vertical dimension was slightly increased (2 mm) in order to restore the existing tooth wear. A full waxup was performed on the mounted study casts (Figs 10 and 11).

The treatment planning for this patient was as follows:

- Initial periodontal treatment (phase 1)
- Extraction of tooth 1 (18) and microscopic examination to verify the clinical diagnosis
- Preparation of maxillary and mandibular posterior teeth for complete-coverage crowns (chamfer margin), and provisional restorations (2 to 5 [14 to 17], 12 to 15 [24 to 27], 21 to 18 [34 to 37], and 28 to 31 [44 to 47]), in increased vertical dimension
- Crown lengthening on the abovementioned regions
- Final preparation for complete-coverage all-ceramic crowns on all the posterior teeth and on teeth 22 to 27 (33 to 43)





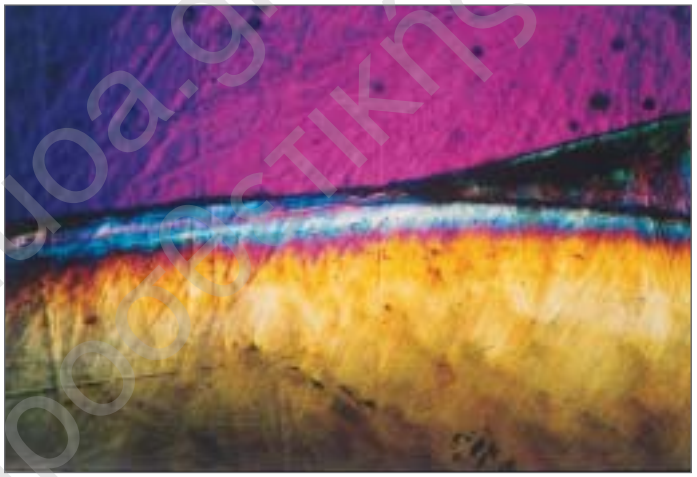
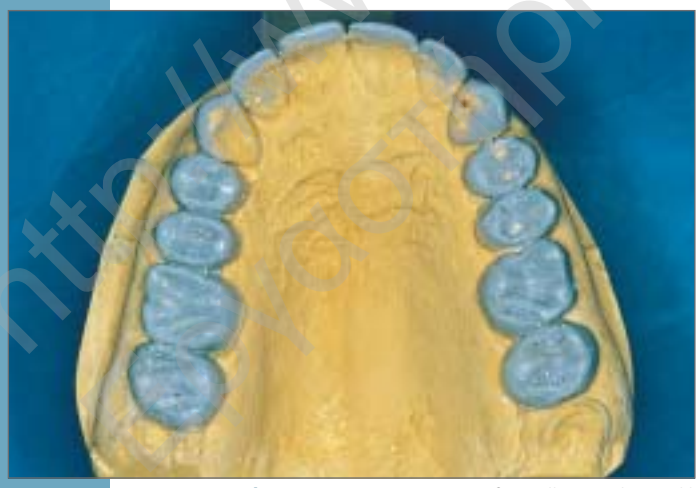
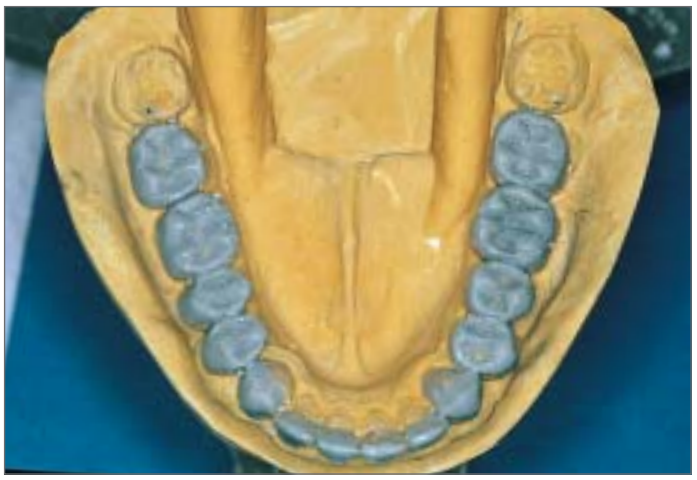


Fig 7 *(above)* Microscopic examination of tooth 1 (18) under polarized light: Hypoplastic enamel layer and an erosion on the labial surface (magnification ×50).
Fig 8 *(top right)* Microscopic examination of tooth 1 (18) under polarized light: Hypoplastic enamel layer thinner than normal, irregular cementoenamel junction with anomalies (magnification ×120).
Fig 9 *(bottom right)* Microscopic examination of the cementoenamel junction on a physiological tooth (magnification ×120).





Figs 10 and 11 Diagnostic waxup of maxillary and mandibular teeth.

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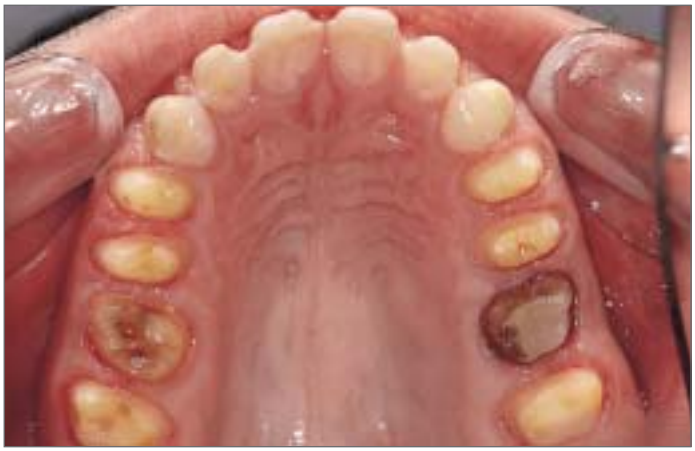


Fig 12 Preparations of the maxillary teeth, occlusal view.

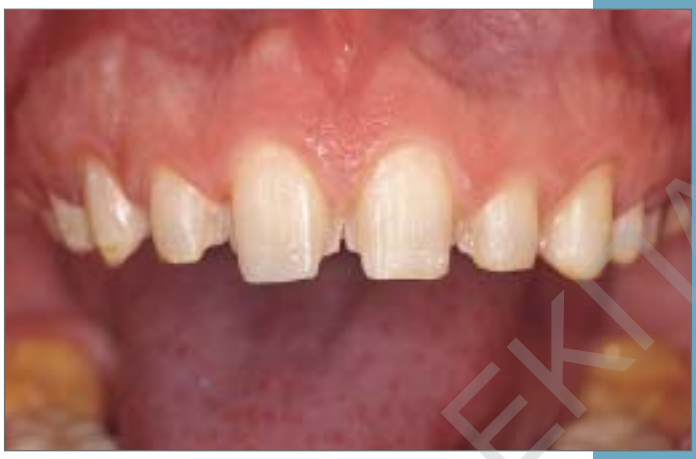


Fig 13 Preparation of the maxillary anterior teeth for laminate veneers.

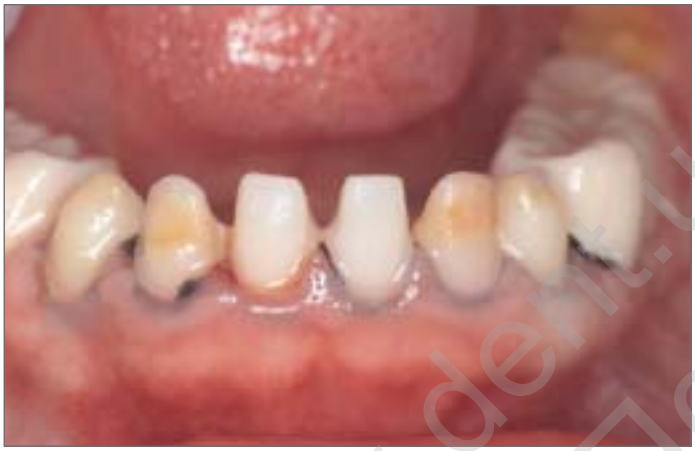


Fig 14 Preparation of the mandibular anterior teeth for complete-coverage crowns. A retraction cord has been inserted in the sulcus.

- Preparation of teeth 6 to 11 (13 to 23) for all-ceramic laminate veneers
- Fabrication of all-ceramic crowns and veneers
- Adhesive cementation of all-ceramic crowns and veneers
- Fabrication of a centric relation splint as a night guard

Treatment steps

After initial periodontal treatment, teeth 2 to 5 (14 to 17), 12 to 15 (24 to 27), 21 to 18 (34 to 37), and 28 to 31 (44 to 47) were prepared for complete-coverage crowns and provisional restorations were constructed chairside with acrylic resin (Dentalon Plus, Kulzer). These splinted provisional crowns were fabricated with a thermopressed transparent sheet (Omnivac) from the waxup.

The enamel was hard and did not show any sign of detachment during preparation. The preparation margins were at the height of the gingival crest. Crown lengthening was performed on all the abovementioned regions, and 2 to 3 mm of crown height was gained. Three months after the periodontal surgery, the teeth were finally prepared with deep chamfer intrasulcular margins (Figs 12 to 14). In order to avoid traumatization of the sulcus depth, a thin retraction cord (000, Ultrapack, Ultradent) was inserted in the sulcus prior to preparation. The same technique was followed for the preparation of the mandibular anterior teeth, 22 to 27 (33 to 43). The preparation depth was controlled with the thermopressed sheet, which was used for the construction of the provisional crowns from the waxup.

The maxillary anterior teeth showed only slight and shallow enamel defects limited to the incisal third area. For this reason, they were restored with laminate veneers. The removal of the surface enamel layer revealed enamel that was normal in hardness and appearance. Provisional restorations for teeth 6 to 11 (13 to 23) were made from light-cured composite resin (Charisma, Kulzer) with the Omnivac sheet and were bonded with a drop of unfilled resin (Heliobond, Vivadent) without etching the enamel.

Final impressions were obtained with polyvinyl siloxane elastomer material (Panasil, Kettenbach) using the corrective impression technique. Occlusal registrations were obtained with a hard addition-type silicon material (Occlufast, Zhermark), maintaining the vertical dimension from the provisional restorations. The working casts were mounted on a semi-adjustable articulator (Hanau, Hanau), and the restorations were waxed in full contour (Fig 15). All-ceramic restorations were fabricated with a leucite-reinforced ceramic (Empress, Ivoclar). The staining technique was applied to achieve maximum mechanical strength. The thermopressed ceramic material, Empress, was used because of its accuracy of marginal fit, biocompatibility, and the possibility of constructing the crowns and veneers from the same material.

After the try-in of the ceramic restorations, occlusal reshaping was performed using articulation paper. The ceramic restorations were glazed and the interior surfaces were etched with hydrofluoric acid 10% for 10 minutes (Ceramic Etching Gel, Ivoclar). The enamel surfaces were etched for 40 seconds with orthophosphoric acid 40% (Etching Gel, Vivadent). The appearance of the hard dental tissues after etching was clinically normal, except for the dark color of the dentin.

The restorations were bonded with a dual polymerization resin cement (Vario-Link, Vivadent) (Figs 16 to 18). After cementation of the restorations, a centric relation splint was constructed from acrylic resin and the patient was instructed to wear it at night while sleeping for protection from possible bruxism. A mutually protected occlusal scheme was designed for this patient to allow even distribution of forces during side movements. There were no interferences in the anterior guidance apart from the contacts on the anterior teeth.

The patient was satisfied with the treatment result and is following a strict 6-month recall program. The restorations have been in clinical use for 3 years with no sign of detachment or fracture.

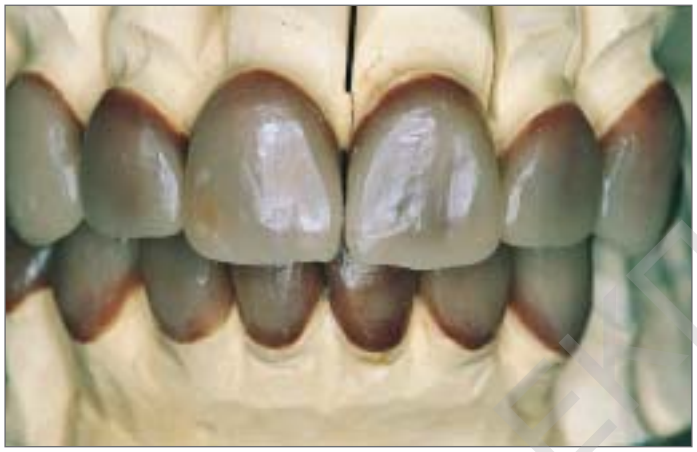


Fig 15 Wax patterns for all-ceramic restorations. Two kinds of wax have been used to achieve good marginal fit.

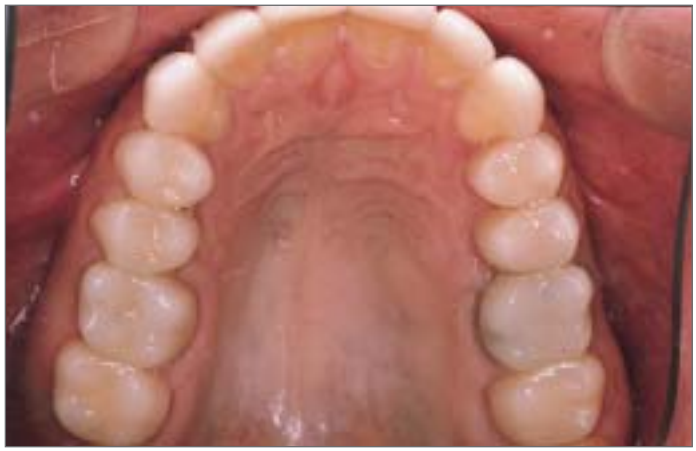
DISCUSSION

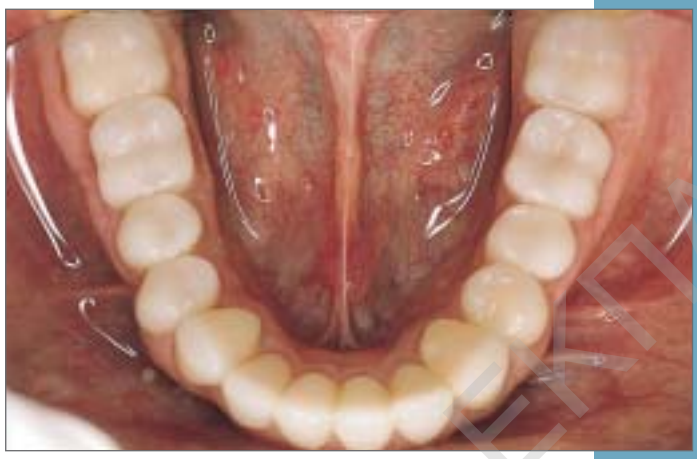
The treatment of a young patient with amelogenesis imperfecta is a challenge for the clinician. Several factors have to be taken under consideration, such as the age of the patient, the quality of the existing dental tissues, the periodontal condition, the pulp-root anomalies, the loss of tooth substance, and the orthodontic condition.

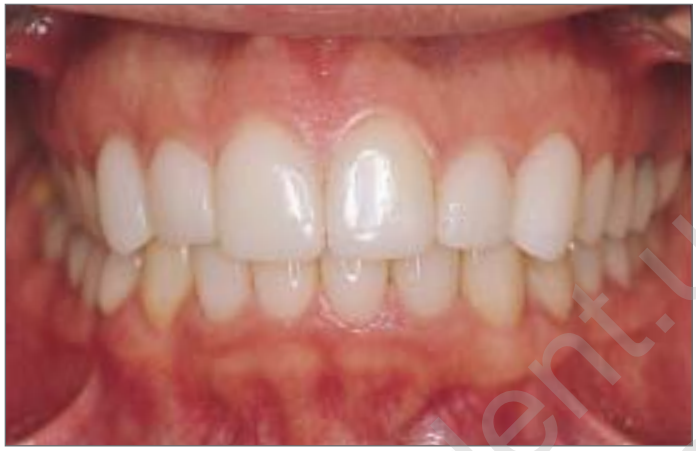
The practitioner has to carefully balance the restorative needs of the patient, the possibility of completing the treatment planning, the protection of the remaining teeth, and the long-term prognosis of the result.

The treatment options vary considerably, depending on the abovementioned factors. Among very young patients or in cases of orthodontic anomalies, the orthodontic treatment is the first step.[23,24] In patients with severe wear on their teeth, an early restorative treatment with prefabricated stainless steel or veneered crowns can improve the esthetics and help the orthodontic treatment.[25]

The prosthodontic treatment usually includes complete-coverage metal-ceramic crowns for functional and esthetic rehabilitation and protection of the remaining teeth.[24,26] The use of all-ceramic bonded crowns (Empress) has also been reported.[27] All-ceramic materials offer certain advantages, including esthetics, marginal fit, and biocom-







Figs 16 to 18 The restorations after bonding with dual polymerization cement. The crown, number 14 (26) appears darker because of the intense discoloration of the underlying tooth.

patibility. A question arises concerning the possibility of bonding partial-coverage restoration (veneers, inlays) on the enamel, although there have been some reports of successful clinical cases.[26,28] In cases of severe hypocalcified amelogenesis imperfecta, the pretreatment with sodium hypoclorite can improve the bonding strength.[29]

The treatment of a patient with dentinogenesis imperfecta with all-ceramic restorations has already been described by the authors.[30] The use of metal onlays in cases of amelogenesis imperfecta to restore the occlusal surfaces and compensate for the reduced vertical dimension has also been reported. Their use, however, was limited to very young patients, and the esthetic result was compromised by the metal surfaces.[31,32]

The decision to keep an enamel layer and use partial-coverage restorations, or remove all enamel and use complete-coverage crowns, depends on the extension and depth of the enamel lesions. The clinical appearance of the enamel during tooth preparation plays a decisive role.

The need for crown lengthening is not uncommon in cases of amelogenesis imperfecta.[26,27] Abrasion and wear on the teeth usually result in reduced crown length. The periodontal condition is usually complicated because of tooth sensitivity and poor oral hygiene.

As in all extensive restorations, the patient should undergo a strict program of scheduled recalls to maintain the achieved result and ensure a good long-term prognosis.

The treatment of patients with amelogenesis imperfecta with overdentures that have been reported has many functional disadvantages and is usually not accepted by the patient.[33,34]

CONCLUSION

The functional and esthetic restoration among patients suffering from amelogenesis imperfecta is a clinical challenge and can be accomplished using a detailed treatment plan. The use of all-ceramic materials offers increased possibilities in the field of esthetics and biocompatibility. This is a very important factor for the patient, who usually has psychosocial problems because of tooth defects from a very young age.

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REFERENCES

1. Cohen MM. Pediatric oral pathology. In: Kelley VC (ed). Practice of Pediatrics. Hagerstown, IL: Harper and Row, 1979:31–65.
2. Forteza S. Amelogenesis imperfecta. Quintessence Int 1980;8:9–19.
3. Darling AI. Some observations on amelogenesis imperfecta and calcification on the dental enamel. Proc R Soc Med 1956;49:39–45.
4. Fritz GW. Amelogenesis imperfecta and multiple impactions. Oral Surg Oral Med Oral Pathol 1981;50: 460–465.
5. De Sort KD. Amelogenesis imperfecta: The genetics, classification and treatment. J Prosthet Dent 1983;49:786–792.
6. Witkop CJ. Hereditary defects in enamel and dentin. Acta Genet Statist Med 1957;7:236–239.
7. Weinnann JP, Svoboda JF, Woods RW. Hereditary disturbances of enamel formation and calcification. J Am Dent Assoc 1945;32:397–418.
8. Chosack A, Eidelmann E, Wisotski I, Cohen T. Amelogenesis imperfecta among Israeli Jews and the description of a new type of local hypoplastic autosomal recessive amelogenesis imperfecta. Oral Surg 1979;47:148–151.
9. Sundell S, Koch G. Hereditary amelogenesis imperfecta. Epidemiology and clinical classification in a Swedish child population. Swed Dent 1985;9: 157–169.
10. Sundell S, Valentin J. Hereditary aspects and classification of hereditary amelogenesis imperfecta. Community Dent Oral Epidemiol 1986;14:211–216.
11. Witkop CJ. Amelogenesis imperfecta, dentinogenesis imperfecta and dentin dysplasia revisited. Problems in classification. J Oral Pathol 1989;17: 547–553.
12. Witkop CJ, Sauk JJ. Heritable defects of enamel. In: Stewart ER, Prescott HG (eds). Oral Facial Genetics. St Louis: Mosby, 1976:151–226.
13. Thakkar NS, Sloan P. Dental manifestation of systemic disease. In: Jones JH, Mason DK (eds). Oral Manifestation of Systemic Disease. 2nd ed. London, New York: Bailliere Tindall Publishing, 1990:480–511.
14. Winter GB, Brook AH. Enamel hypoplasia and anomalies of the enamel. Dent Clin North Am 1975;19: 3–19.
15. Williams SA, Ogden AR. Failure of eruption associated with anomalies of the dentition in siblings. Pediatr Dent 1988;10:130–135.
16. Winter GB. Amelogenesis imperfecta with enamel opacities and taurodontism: An alternative diagnosis for "idiopathic dental fluorosis." Br Dent J 1996; 181:167–172.
17. Aldred MJ, Crawford PJM. Variable expression in amelogenesis imperfecta with taurodontism. J Oral Pathol 1988;17:327–333.
18. Congleton J, Burkes E. Amelogenesis imperfecta with taurodontism. Oral Surg Oral Med Oral Pathol 1979;48:540–544.
19. Person M, Sundell S. Facial morphology and open bite deformity in amelogenesis imperfecta. Acta Odontol Scand 1982;40:135–144.
20. Collins AM, Mauriello SM, Tyndall DA, Wright JT. Dental anomalies associated with amelogenesis imperfecta. A radiographic assessment. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1999;88: 358–364.
21. Emery H, Rimoin DL. Principle and Practice of Medical Genetics. 2nd ed. New York: Churchill-Livingstone, 1991;1:690–696,738–740.
22. Shafer WG, Hine MK, Levy BM. A Textbook of Oral Pathology, 3rd ed. St Louis: Saunders, 1974:48–55.
23. Backmann B, Adolfsson U. Craniofacial structure related to inheritance pattern in amelogenesis imperfecta. Am Orthod Dentofacial Orthop 1994; 105:575–582.
24. Encinas RP, Garcia-Espona I, de Mondelo JMN. Amelogenesis imperfecta: Diagnosis and resolution of a case with hypoplasia and hypocalcification of enamel, dental agenesis and skeletal open bite. Quintessence Int 2001;32:183–189.
25. Rosenblum SH. Restorative and orthodontic treatment of an adolescent patient with amelogenesis imperfecta. Pediatr Dent 1999;21:289–292.
26. Nel JC, Pretorius JA, Weber A, Marais JT. Restoring function and esthetics in a patient with amelogenesis imperfecta. Int J Periodontics Restorative Dent 1997;17:479–483.

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27. Williams WP, Becker LH. Amelogenesis imperfecta: Functional and esthetic restoration of a severely compromised dentition. Quintessence Int 2000;31: 397–403.
28. Rada RE, Hasiakos PS. Current treatment modalities in the conservative restoration of amelogenesis imperfecta: A case report. Quintessence Int 1990;21: 937–942.
29. Venezie RD, Vadiakas G, Christense JR, Wright JL. Enamel pretreatment with sodium hypochlorite to enhance bonding in hypocalcified amelogenesis imperfecta: Case report and SEM analysis. Pediatr Dent 1994;16:433–436.
30. Andritsakis H, Kourtis S, Andritsakis D. All-ceramic restorations for the rehabilitation of a patient with dentinogenesis imperfecta. Quintessence Int 2002;33:656-660.
31. Hunter L, Stone D. Supraoccluding cobalt-chrome onlays in the management of the amelogenesis imperfecta in children: A 2-year report. Quintessence Int 1997;28:15–19.
32. Sengun A, Ozer F. Restoring function and esthetics in a patient with amelogenesis imperfecta: A case report. Quintessence Int 2002;33:199–204.
33. Renner RP, Kleinerman V. Overdenture techniques in the management of oligodontia. A case report. Quintessence Int 1980;11:57–65.
34. Brewer AA, Morrow RM. Overdentures, 2nd ed. St Louis: Mosby, 1980:30–55.

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